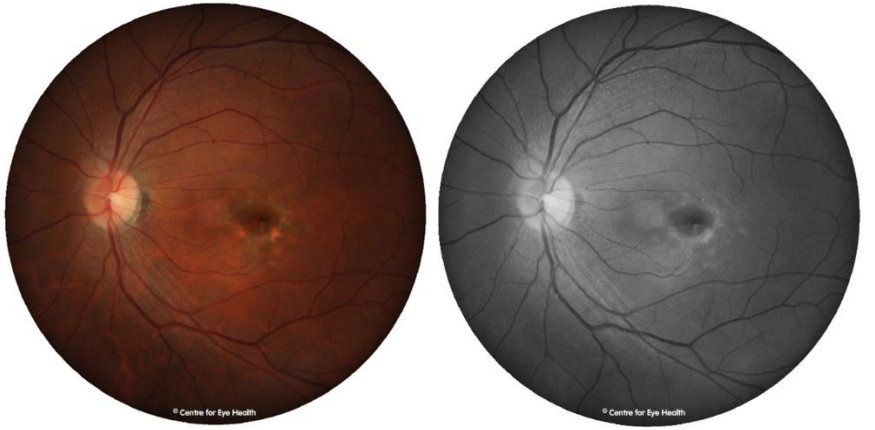
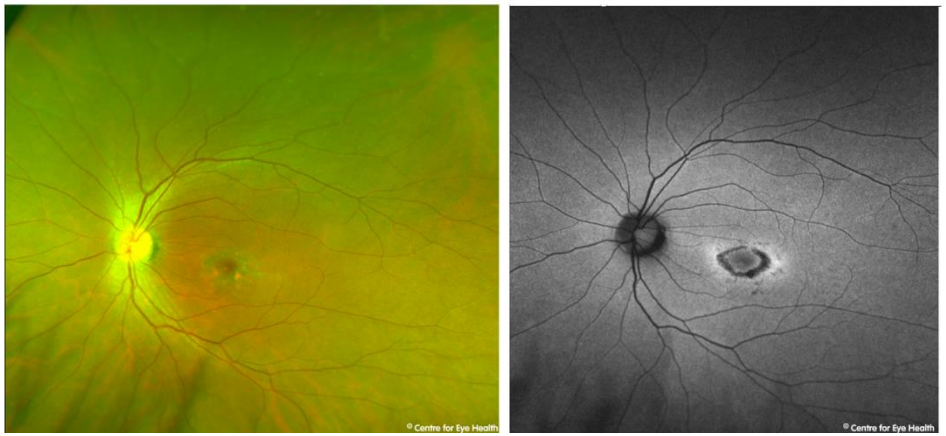
CFEH facebook Cases

CFEH Facebook Case #56

A 43 year old Asian female was referred for investigation of an unusual macular appearance. Both eyes had a similar appearance so only the left eye will be presented here. She reports longstanding "blurry" night vision which caused her to give up driving at night. Her general health is good and she takes no medications. Visual acuity is 6/6-2 in the left eye and Amsler grid shows a central annular area of distortion. Colour vision testing with Lanthony D-15 was borderline and contrast sensitivity below the normal range. What is the likely diagnosis?





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ANSWER

Bull's eye maculopathy.

Depigmentation surrounding the fovea can be seen in the retinal photos. OCT imaging shows a marked thinning of the ONL, ELM, ISe and RPE parafoveally. Autofluorescence imaging showed annular hypo-autofluorescence surrounding the fovea with adjacent hyper-autofluorescence both within and outside the hypofluorescent annulus.

A bull's eye maculopathy is commonly associated with retinal toxicity (chloroquine, hydroxycholoquine, clofazimine) however in this case the patient denied previous use of medications known to cause toxicity. Given this, and the longstanding nature of the night vision problems, the most likely cause is a retinal dystrophy – cone dystrophy, cone-rod dystrophy or Stargardt disease, although the latter is usually associated with retinal flecks so unlikely here.

Referral to a retinal specialist was recommended.